Modern Concepts of Cardiovascular Disease

Published monthly by the AMERICAN HEART ASSOCIATION
44 EAST 23RD STREET, NEW YORK 10, N. Y.

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Vol. XXIV **JANUARY, 1955** **No. 1**

THE MEDICAL MANAGEMENT OF LYMPHEDEMA*

Successful treatment of lymphedema depends on complete cooperation between physician and patient in applying physiological and psychological principles over long periods of time. These require the application of methods which are often uncomfortable and frequently tedious. The physician must maintain optimism, patience, and enthusiasm that may be difficult in the face of slow improvement. At times he will be rewarded by marked, rapid improvement, especially in cases in which the arm is involved. But in lymphedema of the leg, he may have to be content with simply holding the disease in check. In many cases without proper therapy the edema progresses to elephantine size.

CLASSIFICATION

The 1953 edition of the "Nomenclature and Criteria for Diagnosis of Diseases of the Heart and Blood Vessels"[1], defines and classifies lymphedema as follows:

Lymphedema, — This refers to edema resulting from obstruction of the lymphatic flow. Microscopically, the early changes consist of dilatation of the lymphatics and widening of the tissue spaces due to edema. In long-standing lymphedema there is proliferation of the connective tissue, often a variable degree of inflammatory cell infiltration, and fibrotic thickening of the dilated lymphatics.

Clinically, the changes can be correlated with the fact that at first the edematous tissue is compressible ("soft edema"), but later the swollen tissue is firm (so-called "brawny induration" or "hard edema"). It is characteristically pale, in contrast to the cyanosis or rubor seen with venous stasis. The term "elephantiasis" is used for the grotesque deformities of the more severe types. Lymphedema may be "primary" or "secondary" but the former is better designated as "idiopathic".

a. *Primary, or Idiopathic Lymphedema* —

(1) Congenital (Milroy's disease) — This is a hereditary, congenital, non-painful lymphedema involving all or part of one or two extremities. It may be unilateral, involving the upper and lower extremities, and, rarely, additional areas may be involved.

(2) Praecox — This refers to a clinical syndrome affecting young females much more than males. The lymphedema usually appears between the ages of 15 and 35, involving first the feet, later ascending to involve the legs and thighs.

b. *Secondary Lymphedema* — is due to:

(1) Surgical removal of lymph nodes.

(2) Neoplastic invasion of lymph nodes, either by primary or metastatic neoplasm.

(3) Lymphadenitis following

(a) X-ray treatment — Fibrosis and scarring, enhanced by x-ray, may cause obstruction of the lymphatics and resultant edema.

(b) Pyogenic Infection

(c) Granulomata due to (i) Filariasis, (ii) Lymphogranuloma venereum, (iii) Tuberculosis, or (iv) Syphilis.

(4) Dependent edema — Edema of the legs may follow prolonged sitting, as during long trips by auto, rail, or air. It has been reported in persons seated for many hours in air raid shelters. Some complicating factor is usually present, such as girdle that folds and binds the groin when seated, or a chair with a firm edge that presses on the thighs. If the edema is pale, the obstruction is lymphatic. If it is cyanotic, a complicating phlebitis may be present.

CLINICAL OBSERVATIONS

Lymphedema praecox is the most common type of lymphedema of the legs in young persons. It starts most often as a puffiness of the dorsum of the foot in young women. Slowly the puffiness spreads up the leg and thigh. It may be unilateral for many years before involving the other leg.

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At first, the edema disappears during the night when the patient is recumbent. As months go by, it is replaced by a jellylike myxomatous mass.

Constricting garments impede the flow of lymph. Girdles are particularly harmful. They are designed for the upright position. The sitting posture causes them to exert increased pressure and to act as a tourniquet on the lymph and venous flow.

Good lymph flow requires body movement. Sitting for many hours in vehicles, theatres, card-playing sessions, or air raid shelters will frequently result in lymph stasis and edema in an otherwise normal system.

The etiology of lymphedema praecox is completely obscure. From clinical observation it would seem that there is a congenital inefficiency of the lymph system, manifesting itself first where hydrostatic forces are greatest after many years of stress. Kinmonth and Taylor[2] have recently demonstrated (1) the existence of dilated lymph trunks, (2) defective valves, (3) decreased lymph flow, (4) decreased absorption rate of dyes and protein, and (5) a great increase in the concentration of protein in the lymph in lymphedema praecox.

Secondary lymphedema occurs from blockage or extirpation of lymph nodes and channels. It is most frequently seen after radical mastectomy. The arms of such women sometimes reach elephantine size.

Lymphedematous tissue has a decreased resistance to pathogenic micro-organisms. Episodes of acute cellulitis, and erysipelas are common occurrences. Occasionally there is phlebitis. What would be a furuncle in a normal limb may become a major infection. Elevation, moist heat, and appropriate antimicrobial drugs are usually quickly effective against the infection.

DIFFERENTIAL DIAGNOSIS

Because of the frequent episodes of infection in lymphedematous tissue the condition is often confused with thrombophlebitis. The lymphedematous leg is usually pale; pigment deposits are not present; veins are hidden by edema; there is no tenderness except in the presence of infection. The phlebitic leg with edema is cyanotic in dependency; the veins are dilated; collateral veins and plexi are often prominent. Red, hot, tender venous cords are pathognomonic of acute superficial phlebitis. There are deposits of hemosiderin in long-standing venous insufficiency, but not in lymphedema.

TREATMENT

The essence of therapy consists in the judicious application of sound physiological principles.

1. *Gravity:*

Edema of the arms or legs is invariably reduced by elevation[3a]. By reversing the direction of hydrostatic force, the defective lymphatic valves no longer are a deterrent factor. This ideal set of circumstances can be achieved for one-third of every day while the patient sleeps. In cases of leg edema the foot of the bed may be elevated on blocks six inches high. Swollen arms may be suspended in traction. The author[3b] has designed a simple apparatus which accomplishes this in comfort.

2. *Compression:*

While the edema fluid drains out each night, it quickly reaccumulates when the limb is dependent. This accumulation can be prevented by exerting counter pressure. A pressure of 50 mm. of mercury in the foot or hand, decreasing gradually to 15 mm. in the thigh or arm, will accomplish this. Elastic stockings or sleeves, carefully designed and tailored, can be made to do this[3c].

3. *Exercise:*

The pumping action of muscular contraction speeds up lymph flow. With the well-fitted elastic sleeve or stocking in place, full active use of the limb is desirable. Sports, walking, and housework should be encouraged. Standing still or sitting for long periods leads to lymph stagnation.

Walking in deep water or swimming is the single most efficacious method in the treatment of lymphedematous legs. The great hydrostatic force of water pressure, maximal at the foot and gradually less up the leg, is ideal for "milking" the tissue fluids out of the swollen limb. The elastic stocking need not be worn while the patient is in the water.

4. *Dehydration:*

At the onset of therapy a rapid, temporary decrease in swelling can be obtained by the judicious use of dehydration. We employ it during the first week of therapy, usually as a hospital measure. Salt intake is restricted and mercurial diuretics are administered each day. It is of value only if elevation, compression, and the other conservative measures are applied simultaneously.

5. *Massage:*

Effiavage in the direction of lymph flow increases the speed of flow. The patient may be trained to give self-massage. It should be administered for at least ten minutes at the end of the day when swelling is greatest.

6. *Enzyme Therapy:*

The use of an enzyme to hydrolyze hyaluronic acid, a viscous polysaccharide found in the interstices of tissues, has been suggested[4]. The multiple injection of hyaluronidase in varying dosages over a period of time in nine lymphedematous arms and legs, by this author, has failed to improve the swelling. In all instances the edema had been reduced by the above conservative measures prior to the administration of hyaluronidase.

7. *Buried Sutures:*

The use of buried non-absorbable sutures extending from areas of poor lymphatic drainage to normal areas was described by Handley in 1908[5] and revived by Ransohoff[6] in 1945 using nylon thread. In 1951, Zieman[7] described an ingenious method of inserting the sutures through long needles as an office procedure.

A thorough trial by us, in ten patients, has failed. The limbs were first reduced to a minimum size by conservative measures. The sutures were then buried. No further improvement resulted.

COMMENTS ON SURGICAL MEASURES

1. *Sympathectomy:*

We have followed many lymphedematous patients who have had a lumbar sympathectomy performed. Such patients are usually made worse by this procedure.

2. *Ligation of Lymph Channels:*

Incompetent lymph vessels may be ligated in the hope of diverting the lymph stream to collateral lymph channels which have intact valves. One difficulty is in identifying the defective channels. This problem needs further study and experience before comment is justified.

3. *Lymphangiectomy:*

The principle of this operation was described by Charles[8] in 1912. The edematous tissue is removed. The muscles and fascia are covered by skin grafts obtained from the original excised skin of the limb.

This measure is best applied in limbs that have reached gigantic size and in whom the tissue swelling is no longer fluid but has become myxomatous. Pratt[9] has successfully used this treatment for massive lymphedematous legs and Conway[10] has applied the procedure to arms.

PROGNOSIS

The early application of the above physiological principles will prevent the development of massive edema. If the measures are carried out carefully for a period of months, lymphatic efficiency will improve. In many patients the edema will clear completely. In others, improvement may be so marked that some of the therapeutic measures may be dispensed with.

In cases of long standing massive edema the fibrosis that has occurred in the tissue cannot be expected to clear, although the fluid component may be greatly reduced. In such patients the reduction of swelling may be so marked that loose folds of skin are left. The cosmetic appearance will be greatly improved if these folds are excised by an appropriate plastic surgery technique.

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